Saline-agitated echocardiography in diagnosis of cor triatriatum dexter: Case series and literature review

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INTRODUCTION

Saline-agitated transthoracic or transesophageal echocardiography (SATE) is a highly effective diagnostic tool, and is considered the method of choice in showing intracardiac right-to-left shunting, e.g. through the foramen ovale (FO) or in patients with persistent left superior vena cava draining into the left atrium in cases of unroofed coronary sinus. SATE may also be helpful in the visualization of intracardiac flow obstruction with blood rerouting, e.g. in cor triatriatum dexter (CTD) when a prominent Eustachian valve (EV) obturates the tricuspid valve (TV), leading to a pressure increase in the right atrium (RA) and right-to-left shunting *via* the FO.

The chief advantage of SATE is its ability to visualize the direction of microbubbles flow and detect abnormal connections in real-time echocardiography. This is an alternative method to color Doppler flow when multiple artifacts and the lack of an acoustic window can cause difficulties in interpretation and the decision-making process.

MATERIAL AND METHODS

This study included a retrospective analysis of two infants with hemodynamically significant CTD hospitalized in the cardiac surgery department between 2019 and 2023.

The patients were routinely diagnosed with transthoracic echocardiography confirmed in saline-agitated contrast injection, and qualified for cardiac surgery.

Inclusion criteria:

- a prominent EV dividing RA into two chambers with TV obturation or right-to-left shunting *via* FO;
- clinical symptoms including congestive heart failure and/or systemic desaturation;
- saline-agitated injection for CTD confirmation;
- cardiac surgery with membrane excision.

Exclusion criteria:

- a prominent EV in the RA without TV obstruction;
- asymptomatic course without need for SATE or cardiac surgery.

Case 1

A 6-day-old male newborn was referred for cardiac evaluation due to a prenatally diagnosed complex heart defect. The child was delivered at term (40 weeks of gestation) with a body weight of 2550 g. Prenatal ultrasonography showed agenesis of the corpus callosum and aortic atresia, with a large ventricular septal defect and well-developed ventricles. Genetic screening tests showed chromosomal aberrations: Jacobsen and distal trisomy 20q syndromes. On admission, his general condition was poor with symptoms of cardiac compromise and unstable vital signs: tachycardia 160/min, tachypnea with respiratory rate 70/min, mean arterial pressure 40 mm Hg, and SaO_2 of 85%. PGE1 and milrinone infusions were continued. Necrotic enterocolitis was observed which required conservative therapy. Paroxysmal recurrent tachyarrhythmias were registered twice with a heart rate of 250/min and were treated effectively with adenosine. In lab tests, thrombocytopenia (45 000/µl) and increased C-reactive protein (1.79 mg/dl, normal range below 0.5) were found. A blood culture was positive with methicillin-sensitive *Staphylococcus aureus* requiring antibiotics. Transthoracic echocardiography (TTE) showed a critical heart defect with aortic atresia, extremely hypoplastic ascending aorta (2 mm) and aortic arch, large inflow ventricular

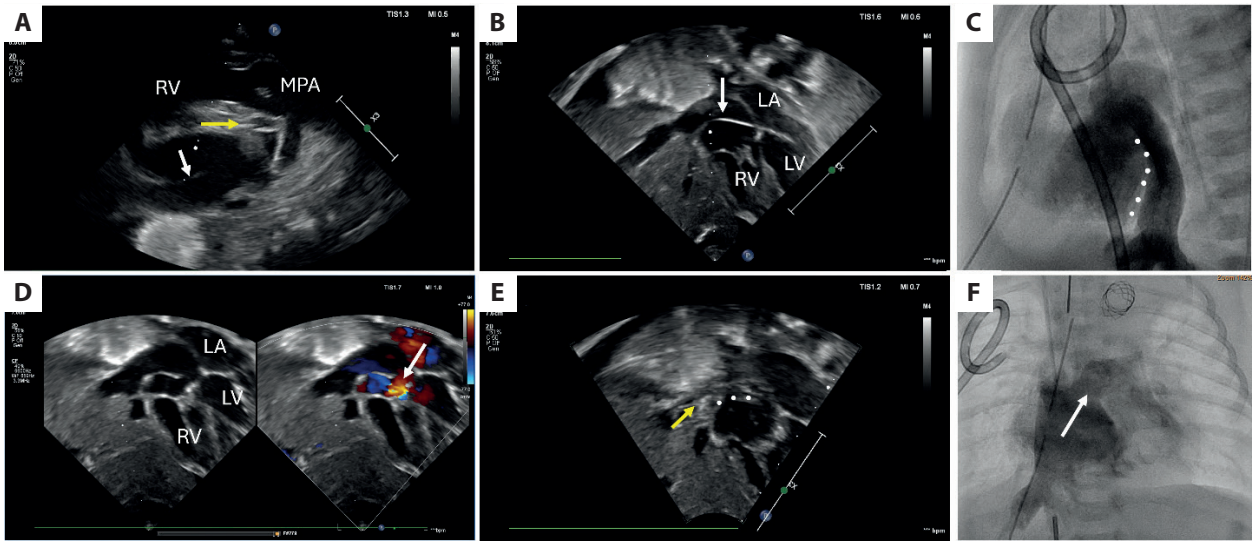

Figure 1. TTE and angiography showing cor triatriatum dexter in a patient with aortic atresia and well-developed two chambers. **A.** TTE parasternal long axis view showing aortic atresia with hypoplastic ascending aorta (yellow arrow), wide pulmonary artery, and prominent EV (white arrow) dividing RA into 2 chambers. **B.** TTE subcostal view showing EV dividing the right chamber into upstream chamber with venous flow and downstream chamber with appendage and tricuspid valve. **C.** IVC angiography in lateral view showing prominent EV (white dots) with division of RA into 2 chambers. **D.** TTE subcostal 4-chamber view with color Doppler as comparison, showing incomplete membrane of EV with small communication and laminar flow. **E.** TTE subcostal view showing saline-agitated inflow with microbubbles (yellow arrow) in upstream chamber. White dots indicate prominent EV. **F.** IVC angiography showing right-to-left shunt *via* atrial septal defect (white arrow)

Abbreviations: EV, Eustachian valve; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle; TTE, transthoracic echocardiography

septal defect, and a prominent EV dividing the RA into upstream and downstream chambers (Figure 1A–B, D; Supplementary material, *Videos S1–S4*). SATE helped to visualize and evaluate the significance of the EV obstruction. The bubbles showed incomplete CTD with preserved flow into the downstream chamber and right-to-left shunt *via* an atrial septal defect (ASD) (Figure 1E; Supplementary material, *Video S5*). Cardiopulmonary compromise required urgent cardiac surgery with a hybrid approach to avoid cross-clamp circulation. Bilateral pulmonary branch bands were established and a Palmaz Genesis 10 mm/19 mm stent was deployed into the arterial duct. Unfortunately, the stent displaced distally. PGE1 infusion was continued, and subsequent heart catheterization was performed with a second Palmaz Genesis 10 mm/19 mm stent deployed into the proximal arterial duct, providing effective systemic flow. Inferior vena cava (IVC) angiography (Supplementary material, *Videos S6–S7*) and hemodynamic evaluation confirmed CTD with a low pressure gradient between the IVC and the downstream chamber due to the large ASD with right-to-left shunt and upstream chamber decompression. At 6 months, Glenn procedure was performed with EV resection. Postoperative hypoxia with superior vena cava syndrome required interventional left pulmonary artery Palmaz Genesis 8 mm/15 mm stent deployment. Pulmonary angiography showed left pulmonary veins stenosis with the need for balloon plasty.

Postoperative course was complicated by progressive cardiopulmonary compromise and hypoxia. Tracheostomy with respiratory therapy relieved the symptoms and provided SaO_2 of above 80%. Following a heart team evaluation, the child was disqualified from additional interventions, and at the age of 6 months was discharged to hospice care.

Case 2

A 21-day-old female newborn was referred to the cardiac surgery department with a diagnosis of CTD. In her medical history, she had been delivered at term in good condition with a body weight of 3580 g. In a pulse oximetry test, low SaO_2 of 88%–92% was found. Her general condition was good, and the physical examination was unremarkable except for mild central cyanosis on the mucous membrane while crying and irregular heart rate on auscultation.

An electrocardiogram and Holter study reported multiple premature atrial beats. TTE confirmed a prominent EV obstructing TV in diastole with right-to-left shunting through the FO (Supplementary material, *Figure S1A–E*, *Videos S8–S10*). During diastole, the membrane became elongated in the shape of a windsock obstructing TV. Pulse wave Doppler showed increased velocity flow (V_{max} — 1.2 m/sec). The FO provided RA decompression with arterial desaturation. Membrane mobility predisposed to atrial premature beats. SATE supported the final diagnosis of CTD, showing the severity of TV obstruction and the bubbles shunting from the RA into the left atrium *via* the FO, and helped to qualify the patient for the operation.

Cardiac surgery in cross-clamp circulation with membrane excision and FO closure relieved the symptoms. The postoperative period was uneventful. TTE showed normal TV inflow and an intact atrial septum, while a Holter study

recorded normal sinus rhythm with single premature beats. At 12 months follow up, her condition was good with normal SaO_2 of >95%.

DISCUSSION

In children with CTD, saline-agitated echocardiography helps to visualize microbubbles outlining the intra-atrial obstruction and right-to-left shunting. During the procedure, two syringes are connected by a three-way stopcock to agitate the saline by exchanging it between the 10 ml syringes. Saline is injected into a peripheral vein in the right upper extremity for FO diagnosis, or into the left upper extremity for left superior vena cava diagnosis. SATE is a valuable diagnostic tool owing its minimally invasive nature. It avoids complications and radiation exposure, and provides real-time TTE assessment of intracardiac anomalies. However, patients with poor acoustic windows may have suboptimal images, risking misinterpretation [1, 2].

Understanding the embryogenesis and hemodynamics of CTD is crucial prior to SATE. During embryogenesis, the sinus venosus collects venous drainage and transforms into the upstream chamber with trabeculated walls, and the downstream chamber with smooth walls, right appendage and TV annulus. CTD results from persistence of the right sinus venosus valve, leading to a triatrial heart with varying degrees of partitioning and hemodynamic disturbances [3].

In very rare cases, CTD may coexist with cor triatriatum sinister, creating quadruple atria [4].

Clinical manifestations depend on the degree of TV obstruction and right-to-left shunting. Most patients, especially adults, are asymptomatic and discovered incidentally. Thereby, due to its asymptomatic nature, complicated cannulation during surgery for other cardiac issues might occur due to fibro-muscular membrane. Besides, longer persistence of abnormal structures may increase the risk of thrombus formation and result in a pulmonary embolism [5]. In cases of long-lasting or significant TV obstruction, easy fatigability, dyspnea, hepatomegaly or even peripheral edema may be observed [6].

Screening TTE may unveil various EV morphologies. In cases of abnormal resorption of the right sinus venosus, a prominent EV may create 3 main types of CTD based on hemodynamics: free-floating EV without significant TV obstruction; incomplete CTD with persistent intra-atrial communication and right-to-left shunting *via* the FO, and most rarely complete CTD with 2 distinct chambers, RV underdevelopment, and right-to-left shunting *via* ASD. Surgical resection of EV can relieve the symptoms of central cyanosis or right ventricular failure.

CTD may present as an isolated anomaly or be associated with other congenital heart defects, as in Patient 1. Differential diagnosis is established using echocardiography, heart catheterization with angiography, cardiac magnetic resonance imaging, or computed tomography to delineate the anatomy of the EV [6].

In conclusion, SATE is an extremely useful tool in the diagnosis of CTD with evaluation of TV obstruction, intracardiac shunts, and IVC congestion. It provides clear visualization and hemodynamic assessment which is crucial in the real-time decision-making process.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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